Abdominal aortic aneurysm with arteritis in ankylosing spondylitis

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Abdominal aortic aneurysm with arteritis in ankylosing spondylitis is described. An abdominal aortic aneurysm, 48-mm in diameter, in a 68-year-old woman with HLA-B27–associated ankylosing spondylitis was successfully replaced with a tube graft. The suture lines of the aortic wall were reinforced with Teflon felt strips. Pathologic examination of the aneurysmal wall revealed hyalinization of the connective tissue, with numerous lymphocytic infiltrates, remarkable calcification, and no elastic fibers. The original structure of the arterial wall was not recognized. These findings are compatible with aortitis reported in ankylosing spondylitis. (J Vasc Surg 2003;38:613-6.)

Ankylosing spondylitis is one of the spondylarthropathies, with distinctive features including a propensity for axial and peripheral arthritis; inflammation at tendinous, ligamentous, or fascial insertions; and a familial pattern of inheritance based on the presence of the class I major histocompatibility complex antigen HLA-B27. In patients with ankylosing spondylitis the incidence of aortic insufficiency is 10% and the incidence of heart block is 5%.[1] Although other cardiovascular manifestations are uncommon, there are reports of polyarteritis nodosa,[2] Takayasu arteritis,[3] aneurysm of the ascending aorta,[4] aneurysm of the entire thoracic aorta,[5] aneurysm of the descending thoracic aorta,[6] and ascending aortitis with aneurysm formation.[7] We describe abdominal aortic aneurysm (AAA) with arteritis in ankylosing spondylitis.

CASE REPORT

A 68-year-old woman with intermittent lower abdominal and back pain was referred to our department. She had been given nonsteroidal anti-inflammatory drugs (NSAID) because of the pain. Abdominal x-ray films disclosed characteristic deformation of the lumber vertebrae, so-called bamboo spine (Fig 1A). Computed tomography (CT) scans revealed an AAA, 48 mm in maximum diameter, with contrast material–enhanced and thickened aortic wall along an anterior aspect of the aneurysm (Fig 1, *B* and *C*). Additional cardiovascular lesions, including aortic root dilatation and aortic or mitral regurgitation on an echocardiogram and heart block on an electrocardiogram, were not recognized, except for 75% stenosis of the left anterior descending artery and 90% stenosis of the posterolateral branch on a coronary angiogram. Criteria were met for classification of spondylarthlopathy proposed by Amor et al,[8] including positive HLA-B27. The patient had undergone laparotomy five times, for appendectomy, ovariectomy, and release of repeated ileus due to intestinal adhesions. Preoperative white blood cell count was 7200/μL, erythrocyte sedimentation rate (ESR) was 101 mm/h, and C-reactive protein (CRP) concentration was 3.32 mg/dL. Antinuclear antibody, rheumatoid factor, and syphilis serologic analysis (Venereal Disease Research Laboratory test and *Treponema pallidum* hemagglutination test) yielded negative results. The patient had neither hypercholesterolemia nor family history, but was a smoker and had hypertension, both risk factors for AAA. Our preoperative diagnosis was inflammatory AAA. The aneurysm was successfully replaced with a tube graft via median laparotomy. The peritoneum was tightly adhesive to the anterior wall of the aneurysm, and appeared shiny and pearly gray, with thickened wall (Fig 2, *A*). Intraoperative findings also suggested inflammatory aneurysm. The suture lines of the aortic wall were reinforced with Teflon felt strips. The postoperative course was uneventful, and CRP concentration declined to 0.15 mg/dL. The graft and anastomosis demonstrated no abnormal findings on an abdominal CT scan obtained 1 month after the operation. Both lower abdominal and back pain disappeared postoperatively without NSAID therapy. Pathologic examination of the aneurysmal wall demonstrated hyalinization of the connective tissue, with numerous lymphocytic infiltrates, remarkable calcification, and no elastic fibers (Fig 2, *B-D*). The original structure of the arterial wall was not recognized.

DISCUSSION

Several reports of aortitis, excluding Takayasu arteritis, with aneurysm in ankylosing spondylitis have been published.[4,6,7] Somer and Siltanen[6] demonstrated aneurysm of the descending thoracic aorta, amyloidosis, and renal carcinoma in a 57-year-old man with ankylosing spondylitis. Microscopic examination of the resected aneurysm revealed changes compatible with severe destructive aortitis. Severe destruction, almost complete lack of elastic tissue, and lymphocytic and plasma cell infiltrations were seen in the degenerated and scarred media adventitia. Kawasuji et al[4] reported aortic valve insufficiency and aneurysm dilatation of the ascending aorta associated with ankylosing spondylitis in a 53-year-old man. Histologic study of the aortic wall disclosed intimal proliferation, fragmentation and scarring of the media, and adventitial scarring with infiltration

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Competition of interest: none.

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0741-5214/2003/$30.00 + 0
doi:10.1016/S0741-5214(03)00231-3

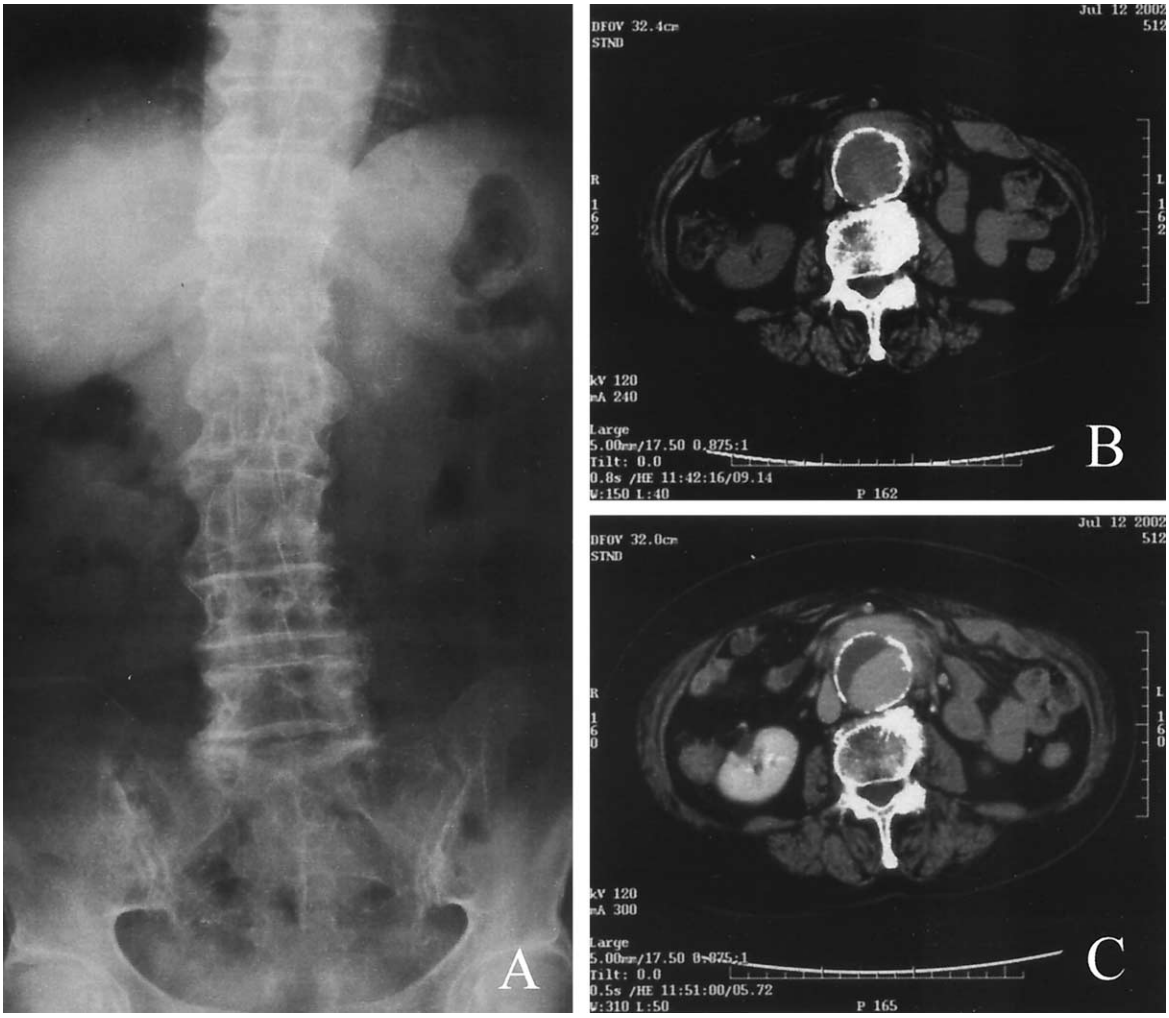

Fig 1. A, Abdominal x-ray film demonstrates characteristic deformation of the lumbar vertebrae, so-called bamboo spine. **B**, Non-contrast-enhanced CT scan reveals an abdominal aortic aneurysm, 48-mm in maximum diameter. **C**, Contrast-enhanced CT scan shows contrast-enhanced, thickened aortic wall along an anterior aspect of the aneurysm.

of lymphocytes and plasma cells into the adventitia and outer layer of the media. Stamp et al[7] reported aortic regurgitation and ascending aortitis with aneurysm formation in HLA-B27–associated spondyloarthropathy in a 24-year-old woman. Histologic analysis of the aorta revealed marked thickening of the intima, with loose fibrous and myxoid tissue, largely destroyed media, and densely fibrotic adventitia. Mild inflammation with lymphocytes was seen around vessels, mainly in the region of the residual media. On the other hand, although Pines et al[9] presented an AAA that simulated ankylosing spondylitis, the pathologic findings were compatible with traumatic aneurysm without signs of degenerative or inflammatory disease.

Bulkley and Roberts[10] reported the characteristic pathologic features of aortitis associated with ankylosing spondylitis in necropsy studies. The aortic inflammatory process was limited to the aortic wall behind and immediately above the sinuses of Valsalva, particularly behind and adjacent to the commissures. Nevertheless, the inflammatory process uncommonly extends more distally into the ascending thoracic aorta,[4,7] descending thoracic aorta,[6] or abdominal aorta.[11] Ansell et al[11] demonstrated ankylosing spondylitis in a 27-year-old man whose aorta showed patchy hyaline thickening to below the renal arteries, especially around the orifices of branches, longitudinal wrinkling, and a few spider scars, closely resembling the changes of syphilis. In the present case, although the histologic features are compatible with aortitis reported in ankylosing spondylitis, they do not appear to be distinct from those seen in inflammatory AAA. Thus it is unclear that the aneurysm is secondary to ankylosing spondylitis, and these may be unrelated but coexisting diseases. The inflammation was located in the abdominal aorta, with aneurysm formation. There was, however, no aortic root dilatation suggestive of inflammation on an echocardiogram. As in giant cell arteritis, polymorphic structures within the HLA molecule

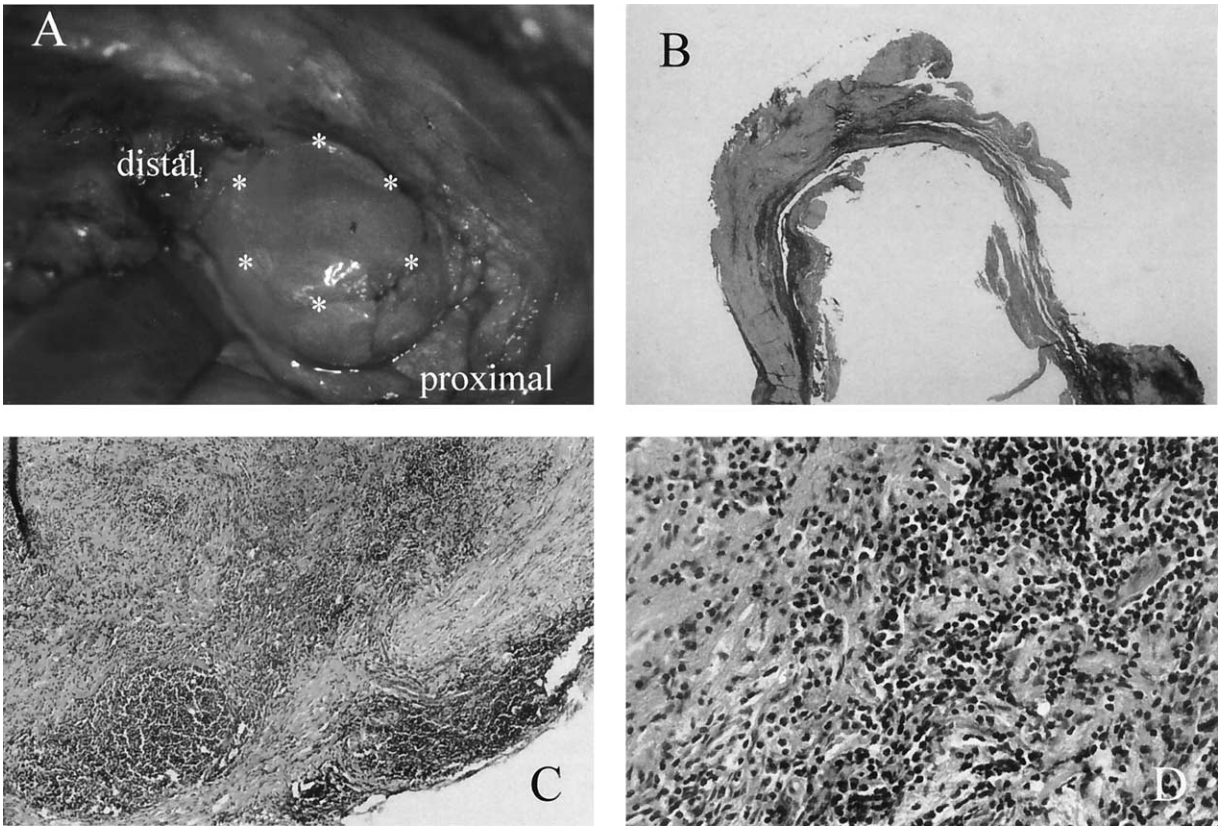

Fig 2. **A**, Peritoneum (surrounded by asterisks) is tightly adhesive to anterior wall of the aneurysm, appearing shiny and pearly gray, with thickened wall. **B**-**D**, Pathologic examination of the aneurysmal wall demonstrated hyalinization of the connective tissue, with numerous lymphocytic infiltrates, remarkable calcification, and no elastic fibers (hematoxylin-eosin; original magnification ×20, ×100, and ×400, respectively).

may predispose to development of inflammatory AAA.[12] On the other hand, HLA-B27 is found in 90% of North American white patients with ankylosing spondylitis and 60% of African Americans with the disease.[13] The relationship between inflammatory AAA and HLA-B27–positive ankylosing spondylitis, however, is unclear.

Surgical treatment of aneurysm with aortitis in ankylosing spondylitis has been successfully performed, and resulted in no anastomotic pseudoaneurysm during short postoperative follow-up.[4,6,7] However, a second operation to repair graft detachment is occasionally required in surgical management of Behçet's aortitis.[14] It is crucial to reduce inflammation preoperatively and postoperatively, to reinforce the suture line, and to carefully select the operative procedure when treating cardiovascular disorders caused by systemic inflammatory disease, eg, Takayasu disease, systemic lupus erythematosus, rheumatoid arthritis, and Behçet's disease.[15] In the patient with HLA-B27–associated spondyloarthropathy and severe ascending aortitis described by Stamp et al,[7] sulfasalazine, prednisone, cyclophosphamide, or azathioprine was administrated until the operation, because of excessive systemic inflammatory response and rapid expansion of the ascending aortic aneurysm. In the present case the suture lines of the aortic wall were reinforced with Teflon felt strips to prevent graft detachment, resulting in anastomotic pseudoaneurysm. Although NSAID had been administered to relieve abdominal and back pain until surgery, systemic inflammatory response, ie, ESR of 101 mm/h and CRP concentration of 3.32 mg/dL, was recognized preoperatively. Immunosuppressive drugs were not administered because the AAA was considered not associated with ankylosing spondylitis but an "inflammatory aneurysm" preoperatively, and the inflammatory response resolved without medication postoperatively.

We thank Emeritus Professor Dr Kiyoshi Inada, Gifu University, for assistance in preparing the manuscript.

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Submitted Nov 12, 2002; accepted Jan 31, 2003.